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IDIOPATHIC RETROPERITONEAL FIBROSIS: A CASE OF UNILATERAL LESION

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Here we report a case of idiopathic retroperitoneal fibrosis, a unilateral case, which resulted in remission by ureterolysis with intraperitonealization. A 24-year-old female was hospitalized because of hydronephrosis. Many examinations were performed with the suspicion of malignant disease, but no apparent cause was detected. We suspected idiopathic retroperitoneal fibrosis and performed surgical exploration. The pathological diagnosis was retroperitoneal fibrosis. Ureterolysis with intraperitonealization of the ureter was performed. Six months later, marked improvement of the right hydronephrosis was observed without steroid therapy.

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Key words: Idiopathic retroperitoneal fibrosis, Ureterolysis, Intraperitonealization

Idiopathic retroperitoneal fibrosis¹⁾, first recognized in 1905 was established as a clinical entity in 1948 by Ormond²⁾. It shows various clinical manifestations, especially ureteral obstruction in the urological field. Here we report a patient with the unilateral involvement of idiopathic retroperitoneal fibrosis who showed remission after ureterolysis with intraperitonealization of the ureter.

CASE REPORT

A 24-year-old female was admitted to the hospital because of intermittent lowback and flank pain, and fever, but otherwise healthy with no significant medical history. Physical examination was normal. Routine urinalysis was unremarkable. Urine cytology showed no malignant findings and the culture was negative. Laboratory data demonstrated a hemoglobin of 8.7g/dl, red blood cell of $325 \times 10^3/\text{mm}^3$ and white blood cell of $6,200/\text{mm}^3$, with a normal differential. The erythrocyte sedimentation rate was 88 mm/hr (normal, <15 mm/hr). The serum sodium was 138 mEq/l; the potassi-

um, 3.7 mEq/l; chloride, 107 mEq/l. BUN and Cr were 17 mg/dl and 0.6 mg/dl, respectively. Total protein was 6.6 g/dl, and A/G ratio was 0.89 (normal, 1.65~2.65). An elevated IgG of 2,211 mg/dl (normal, 980~2,070 mg/dl) was detected. CRP was 5.95 mg/dl (normal, <0.3 mg/dl). A serological study revealed a positive antinuclear antibody with a titer of 1:80, a negative anti-smooth muscle antibody and anti-mitochondrial antibody. The serum components C3 and C4 were 93 mg/dl (normal, 45~80 mg/dl), 25 mg/dl, respectively. An IVP showed hydronephrosis on only the right side. DIP demonstrated that the right ureter was deviated abruptly medially at the level of L5 with fusiform narrowing at the distal ureter (Fig. 1). Cystoscopic findings were unremarkable. Retrograde pyelography of the right ureter revealed dilatation of the caliceal system with an obstruction near the iliosacral joint (Fig. 2). Computed tomography demonstrated the dilatation of pelvis and ureter on the right side, but no remarkable mass was observed near the stenotic

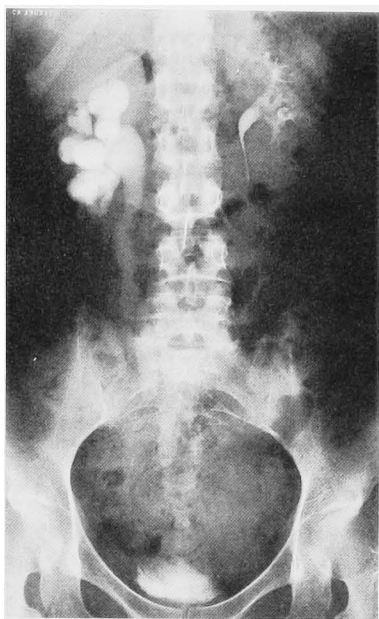


Fig. 1. DIP demonstrated that the ureter was displaced medially at L5 with stenosis at the iliosacral joint.



Fig. 2. Retrograde pyelography: the stenosis was observed at iliosacral joint.

area of the ureter. MRI also showed a similar finding. Gastrointestinal examina-

tion was unremarkable. Ga scintigraphy showed no remarkable accumulation. To exclude the possibility of endometriosis, laparoscopy was done. No evidence of endometriosis was detected and grayish white, hard tissue was observed around the area where the distal ureter seemed to go through. Surgical exploration was carried out on Mar. 26, 1992. The entire right ureter was encased in fibrous tissue with no evidence of tumor at the level of obstruction. Ureterolysis of the right ureter was done and then, the ureter was placed intraperitoneally. The pathological examination revealed that the periureteral tissue was fibrous with unremarkable inflammation. The postoperative course was uneventful without steroid therapy. Six months later, an IVP showed marked improvement of the hydronephrosis and the patient remains asymptomatic (Fig. 3).

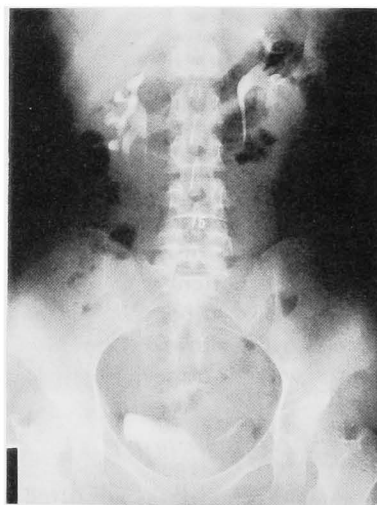


Fig. 3. Six month later, IVP showed marked improvement of the hydronephrosis.

DISCUSSION

The etiology of retroperitoneal fibrosis remains unclear^{3,4)}. Clinical diagnosis of idiopathic retroperitoneal fibrosis is difficult⁵⁾. Before diagnosing this disease, all identifiable causes should be excluded; i) malignancy, ii) retroperitoneal injury, iii) infectious agents, and iv) drugs. In our case, all causes described above were ex-

cluded. By laparoscopic examination, the area around the distal ureter appeared grayish white, hard, and fibrous plaque. Although the definite diagnosis was not clear, this procedure was considered to be very helpful to exclude all unidentifiable causes and to make a diagnosis prior to surgery. However, the final diagnosis should be made at the time of surgical treatment.

The treatment of idiopathic retroperitoneal fibrosis was divided into two categories. One is steroid medication. The ureteral obstruction has been reported to be resolved by steroid therapy without operation^{6,8)}. However, care must be taken, since the individual response to therapy would obviously depend on the degree of acute inflammation versus hyalinized fibrosis⁹⁾. Recently, azathioprine, an immunosuppressive drug is recommended for the cases of idiopathic retroperitoneal fibrosis¹⁰⁾ recurrent and resistant to steroid therapy. The other is surgical treatment. Generally ureterolysis is performed with or without intraperitonealization. Wagenknecht et al.¹¹⁾ reported that 68% of patients recurred after only ureterolysis. On the other hand, combined with intraperitonealization, the rate of recurrence was reduced to 14~56%. Surgery followed by steroid therapy is, has been reported to be in general, preferable to ureterolysis alone^{12,13)}. However, the best strategies for the treatment is still controversial. Since, in our case, the sclerosing phase of the tissue indicated that steroid therapy would not be effective, ureterolysis with intraperitonealization alone was performed, which was very effective without steroid therapy.

As shown in this case, 45% of the patients have been reported to have unilateral involvement; in 23% of them there was progression to the contralateral side during follow-up^{14,15)}. Therefore careful follow-up by CT¹⁶⁾ and MRI¹⁷⁾ is necessary and important besides laboratory data, although the best method of monitoring disease remain obscure.

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和文抄録

特発性後腹膜線維症の1例

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24歳，女性．側腹部痛を主訴として受診．IVP にて右水腎症を認めた．DIP にて L5 付近において尿管が内方に偏位し，その遠位において狭窄を認めた．逆行性腎盂造影では仙腸関節付近に尿管の狭窄を認めた．CT, MRI では明らかな腫瘤を認めなかった．悪性病変をふくめ種々の原因検索を行ったが明らかな

所見をえられなかったため，特発性後腹膜線維症を疑い試験開腹を行った．病理組織学的に後腹膜線維症であった．尿管剝離術および腹腔内留置術を施行した．6ヵ月後，水腎症は著明に改善し現在のところ再発の兆候を認めない．

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